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A Case of Symmetrical Cutaneous Atrophy of the Extremities

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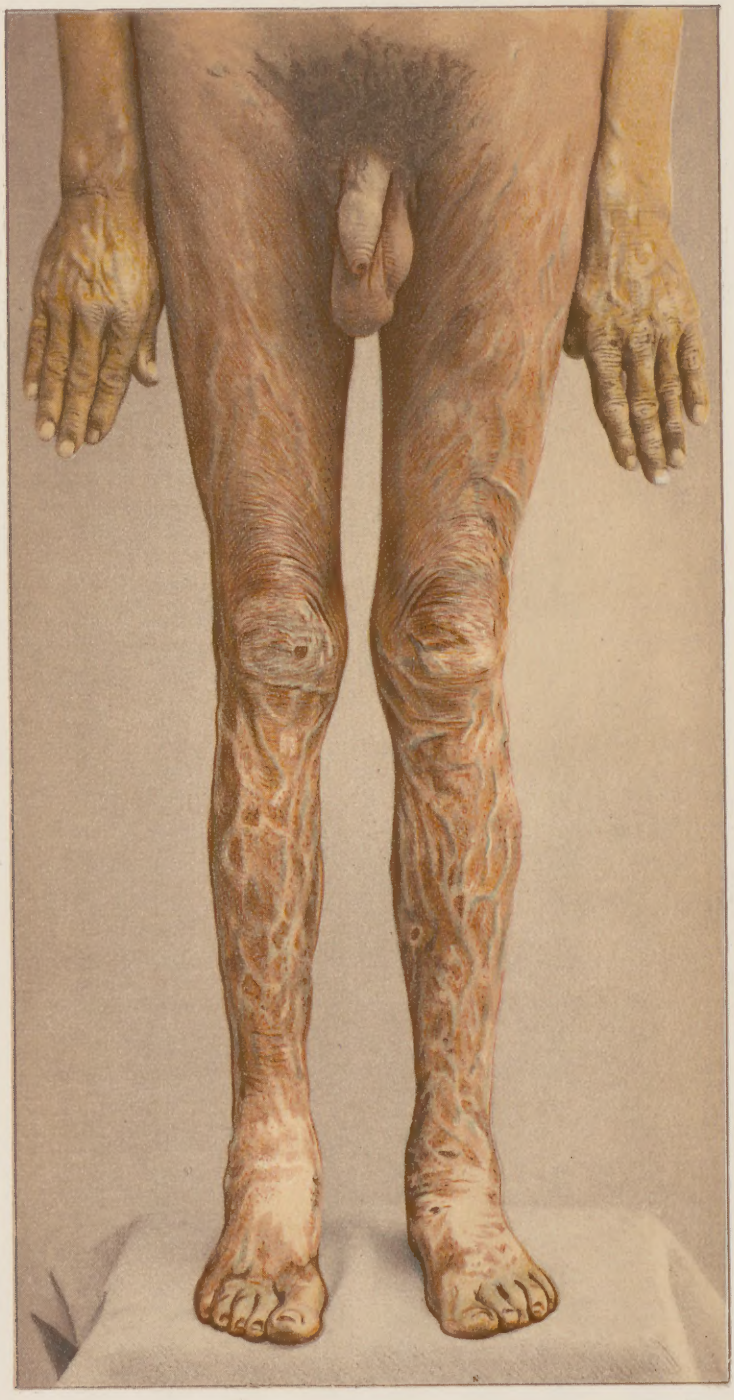

DR. BRONSON'S CASE OF
SYMMETRICAL CUTANEOUS ATROPHY OF THE EXTREMITIES.

A CASE OF SYMMETRICAL CUTANEOUS ATROPHY OF THE EXTREMITIES.*

By EDWARD BENNET BRONSON, M. D.,
Professor of Dermatology in the New York Polyclinic.

MR. PRESIDENT AND GENTLEMEN:

The case which I am about to report, while not a unique one, is nevertheless such a rare form of atrophy of the skin that it has seemed to me not unworthy of engaging your attention. The patient exhibiting the peculiar affection was sent to me at the Polyclinic by Dr. F. P. Griswold, of Meriden, Conn., about the middle of last October. I was then only able to make a few notes and secure some indifferent photographs before the patient returned to his home. A second visit was paid me at my request about one month ago. At this time I could not see that any changes had taken place in the appearances of the disease during the six months' interval. Apparently the case had remained quite stationary. In the presence of the patient on this occasion two of the photographs taken previously were colored. I regret that my opportunities for studying the case have been so meager. Such notes as I was able to make are as follows:

The patient is a man forty-five years of age, native of Germany, by occupation a varnisher in brass and iron works. His appearance is that of a well-developed, well-nourished person, and his manner indicates fair intelligence. He is of spare, sinewy build and of brownish complexion, with dark hair and eyes. He states that his mother died

* Read before the American Dermatological Association, May 31, 1894.

of "brain disease," and an aunt, it would seem, has some mild form of paranoia. In other respects the family history is good. The patient formerly drank rather freely, but has generally led a regular, hard-working life, is married, and has three healthy children. He gives no evidence of syphilis nor of arthritic disease, and his first and only serious ailment, he states, is the skin trouble with which he is now affected. The first sign of this was noticed fourteen years ago, in the region of the left ankle. The skin at this place seemed thinner and more sensitive, and the blue veins began to show prominently. There was also some scaling of the skin, and from time to time rather dry sores appeared over the bony prominences and were difficult to heal. Little by little similar changes began to appear higher up. A year or two later the same thing was noticed upon the right leg. These changes, it would seem, were neither preceded nor accompanied with any form of inflammation excepting such as attended the ulcers or sores, and these were of a very indolent character. Nor had there been any previous disease or injury to which the affection could be ascribed. It was apparently spontaneous and idiopathic. In course of time the hands and arms became similarly affected, but not, the patient thinks, till about five years ago. As the disease progressed and extended higher, it has been attended with occasional pain or discomfort, which thus far has affected only the lower extremities. The chief thing complained of has been a sense of great fatigue after work and long standing. This lasts, he says, for an hour or two after his work is over, and then disappears. So far it does not seem to have been sufficient to seriously interfere with or curtail his working hours, but chiefly because of the apprehension that it would come to this was he led to seek medical aid. Besides this feeling of fatigue, there have occasionally been sharp, shooting pains that would come in little shocks, especially at night, and often in the soles of the feet. There has been some itching in the vicinity of the ulcers that have developed from time to time about the ankles and anterior surfaces of the legs. No other abnormal sensations, such as a sense of constriction, tingling, numbness, "pins and needles," heat or coldness, have been complained of. It was noticed, however, that the affected surfaces were abnormally sensitive to touch, though not particularly to changes of temperature.

Status præsens.—When the patient is stripped a marked contrast is presented between the skin of the legs from the hips down, together with the lower two thirds of the arms, on the one hand and the surface of the rest of the body on the other, which latter everywhere appears perfectly normal. By comparison with the trunk the extremities ap-

pear thin, as if slightly shrunken; they are darker in color, show the courses of the veins more clearly, and are covered with multitudes of wrinkles diversified with smooth brownish or white interspaces that have a glazed appearance, resembling, but for the dusky color, a crumpled sheet of waxed paper or gold-beater's skin. (See colored plate.) The wrinkling for the most part follows the cleavage lines of the skin. It is most marked about the extensor aspects of the knees and wrists, especially the former. About the ankles and lower part of the legs the skin is a little scaly.

The extent of the disease on the lower extremities is about the same on either side. It begins below at a pretty sharply defined line, about an inch and a half above the soles at the side and back of the feet, and one inch back of the roots of the toes in front, whence the affection extends upward continuously, embracing the entire circumference and length of the legs; in front to a point within two or three inches of the flexure of the thigh; at the sides is bounded by a line that curves over the trochanter and behind reaches above the nates and half way up the surface of the sacrum. The cleft of the

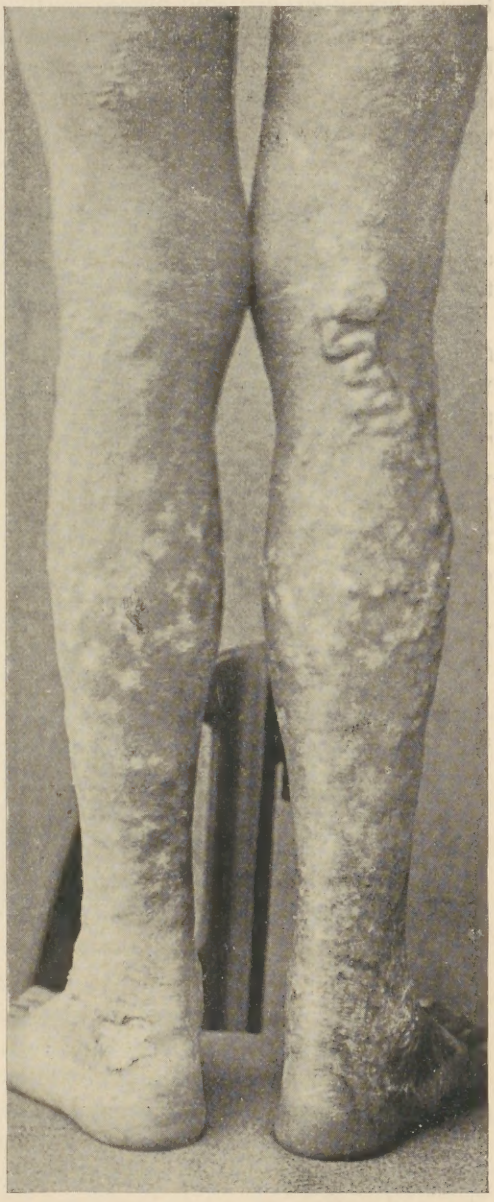

FIG. 1.

nates, the perinæum, and genitals remain unaffected. The line of definition above is not very clear, though there is a marked contrast between the normal white velvety skin above and the reddish purplish brown, wrinkled, dry, and parchmentlike surfaces below.

Upon the upper extremities the skin is affected from the base of the fingers (which latter are not affected) posteriorly and the palms anteriorly to an oblique line encircling the arm a little above the elbow. Its highest point, which on both sides is at the back of the arm, is near the junction of the lower and middle thirds, and is a trifle higher on the left side than on the right. Here, as on the lower extremities, while there is a sufficiently marked contrast between the atrophied and the sound skin, the line of definition is somewhat indistinct.

The Color.—This appears to be due to a blending of purple, red, and brown. (See colored plate.) Those elements of the skin upon which its opacity and consequent whiteness depend seem to have disappeared, permitting the blood-vessels underneath to show through. This is especially evident near the margins of the atrophied portions, where veins which are clearly perceptible in the latter are suddenly lost to view as they enter the area of normal skin. Besides the purple hue of the numerous veins there is a red reflex from the arterioles and capillaries. In some places there is a lilac coloring due to the combined effect of arteries and veins. Almost everywhere also there is a brownish discoloration which is made more evident when pressure is used so as to produce a temporary ischæmia of the part. In many places this brown pigmentation is seen to be punctate or in small lentigolike spots. It occurs over almost the entire atrophic area, and contributes a considerable quota to the general dusky discoloration. The color varies, however, with the patient's posture. Especially in the legs after the patient has been standing for a length of time the veins become turgid and greatly dilated, standing out as prominent tortuous ridges, and their deep purple color becomes everywhere predominant, giving the appearance of cyanosis.

In certain places, more especially over the legs and on the backs of the feet and hands, there are numerous whitish, scarlike patches where the atrophy has been more profound, and perhaps in some places corresponding to the sites of old ulcerations. (Fig. 1.) Even in many of these scarlike patches the punctate brown spots are distinctly present. Aside from these scarlike patches there are scattered over all the atrophic regions a multitude of small, slightly depressed spots which are apparent only on closer inspection, and which seem to imply that the degenerative process has not been absolutely uniform, but has been

more pronounced in certain places than others. Such shallow depressions are scattered abundantly over the thighs, buttocks, and arms.

Near the outer malleoli of both legs are shallow, indolent ulcers with sharp-cut adherent edges and gray base, devoid of granulations and showing a scanty serous secretion.

The wrinkling of the skin is apparent almost everywhere in the affected regions, though less marked on the legs than on the thighs and buttocks. In most places the wrinkles are extremely fine, looking at

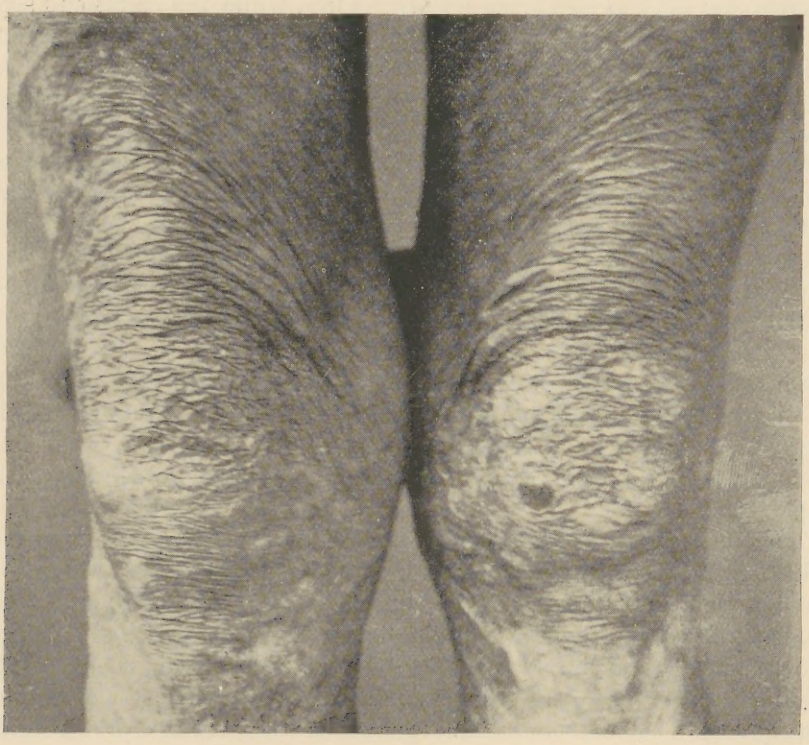

Fig. 2.

a little distance like minute striæ. About the knees they are most pronounced. (See Fig. 2.) Here the thinned skin is thrown into numberless parallel transverse folds curving above and below the patellæ, leaving between them flat, smooth, glistening surfaces that look as if waxed or varnished. These flat, smooth surfaces are most marked over the patellæ. The wrinkling is well marked also over the nates, especially near the folds between the nates and thighs. The

lines curve gracefully from the outer aspect of the thigh around the nates toward the cleft. Just back of the toes the skin is thrown into fine wrinkles or striations that radiate outward from the atrophied portion, which here is whitish and scarlike with a lilac-colored border.

The hairy growth has almost entirely disappeared from the affected region, including the lanugo as well as the coarser hairs. Apparently the little dots of pigment scattered over the legs and arms mark the sites of degenerated hair follicles.

No sweating is apparent in the areas of atrophy. The patient states that his legs and arms are always dry, while the palms and soles sweat freely. While examining him, it was noticed that all around the borders of the feet, just without the atrophied portion, the skin was decidedly moist, while just above it was perfectly dry. To the touch the skin was like dry parchment. There seemed to be no change of temperature. It was neither below nor above the normal. Almost everywhere the skin was freely movable over the subjacent structures. On the legs, however, it was less so than elsewhere, and in places here was adherent. Elsewhere it could be readily pinched up, always in very thin folds, however, which on being released returned again to the niveau, though a little more slowly than in the normal skin.

The sense of touch over the affected region is very little if any diminished, but the skin is distinctly hyperæsthetic. The patient shrinks away from the points of the æsthesiometer much more when the latter touch the atrophied skin than when the sound skin is touched, due evidently to loss or thinning of the protective layers of the epidermis.

So far as could be ascertained, the organic muscles of the extremities were not noticeably atrophic. The patient was not aware of any weakness of the limbs, and the muscles were firm and hard to the touch. The abnormal sense of fatigue, however, of which the patient complained, would imply that they were not entirely unaffected. The thinner appearance of the legs is doubtless, chiefly at least, attributable to the loss of subcutaneous fat. The muscular strength of the limbs seems little if any impaired, and apparently there is perfect co-ordination as well as normal tendon reflexes.

The above, though evidently a case of idiopathic atrophy of the skin, differs very materially from all the common forms of macular, striate, or diffuse atrophia cutis propria. The latter, as a rule, are simply of the quantitative type, and resemble merely superficial cicatrices. Still less does this case resemble the stationary form of the xeroderma of Kaposi. Nor does it correspond to the symptomatic atrophies, such, for example, as succeed scleroderma and morphœa. Not only is there

the history in such cases of a precedent condition of infiltration or induration, with more or less stiffness of the skin, but rarely even in the atrophied stage is the affected area devoid of a certain degree of condensation and immobility. Nevertheless, in their ultimate state they may come to resemble the case which I have described above, and such cases have occasionally been reported as cases of idiopathic atrophy. In this category should, in all probability, be classed the "general iodiopathic cutaneous atrophy" of Wilson, as well as the cases of Schwimmer* and Glax,† though these latter are both improperly referred to the xeroderma of Kaposi, that of Schwimmer being also denominated "atrophia cutis universalis."

A case reported by Judassohn before the German Dermatological Society in September, 1891,‡ under the name of atrophia maculosa cutis, bore a certain resemblance in some of its features to the one just described. The lesions were symmetrical, and in some of the affected places the skin had the same dusky-red color and translucency, and was inelastic, wrinkled, and loosely adherent, but the atrophied spots were preceded by an eruption of papular elevations, that remained circumscribed and discrete, varying in size from that of a lentil to that of a silver mark piece. On account of the looseness and inelasticity of the skin, together with the preceding papulo-erythematous condition, he proposed the name *anetodermia erythematodes*. In a similar case presented by Beer# at the Vienna Dermatological Society in February, 1892, the lesions were preceded by pronounced œdema. In both of these cases the atrophy affected chiefly the connective tissue, and more particularly the elastic fibers.

The clinical type to which my case most clearly corresponds was first described by Buchwald,‖ and so peculiar is the type and well differentiated is it by this writer that it might well be known as *Buchwald's atrophy*. In his case the affection occurred in a man thirty-six years of age, and had existed for sixteen years. It began at both knees and extended at first both upward and downward, later the extension being only upward, but upon the legs below the knees indolent ulcers often formed. The greatest extent was reached in one year from the inception of the disease, though within the affected areas the degeneration continued to increase. There were no prodromal symptoms, and

* *Die neuropathische Dermatosen*, Wien, 1883, p. 189.

† *Viertelj. f. Derm. u. Syph.*, 1875, p. 114.

‡ *Monatshft. f. p. Derm.*, 1892, p. 621.

Arch. f. Derm. u. Syph., 1892, p. 835.

‖ Ein Fall von diffuser idiopathischer Haut-Atrophie. *Viertelj. f. Derm. u. Syph.*, 1883,, p. 543.

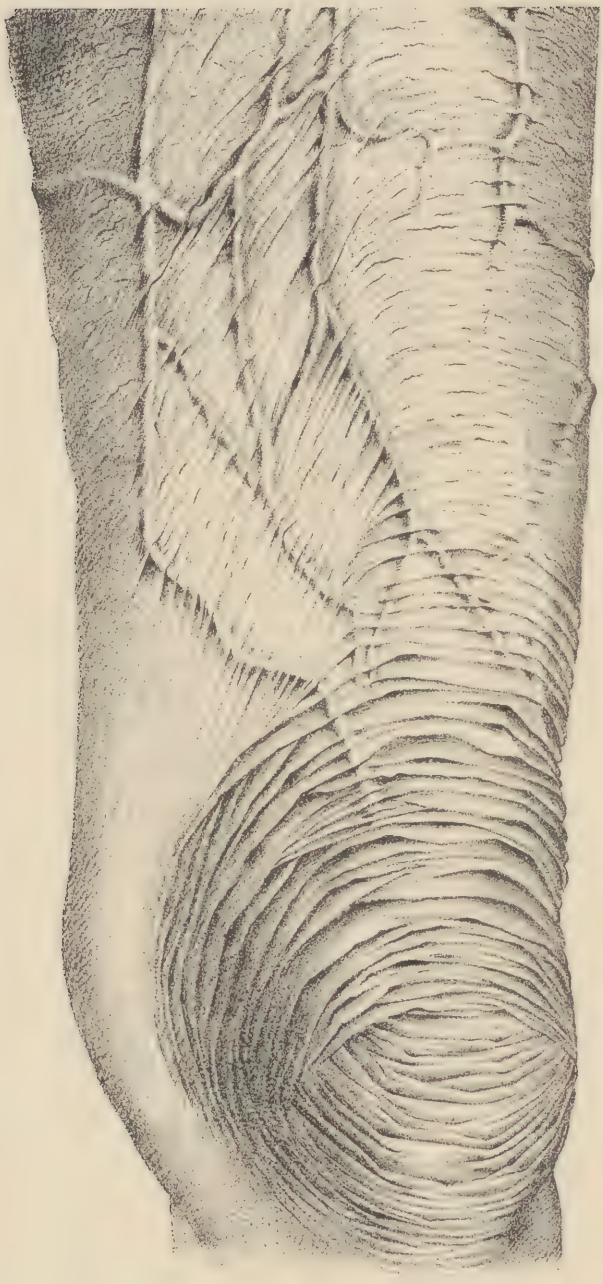

FIG. 3.—Buchwald's Case.

the ætiology of the case was as obscure as that of mine. The entire surface of both knees and thighs was involved, reaching in front to within five centimetres of the inguinal fold, and behind extending up over the nates, much as in my own case. (Fig. 3.) The limits of the disease above were rather abrupt, the division between the normal, well-nourished, and fat skin above, and the somewhat sunken, darker-colored areas of atrophied skin below being marked by a pretty distinct ridge. The affected skin was freely movable over the subjacent structures, was nowhere bound down, rigid, nor œdematous. It could be raised in large folds which, when released, disappeared rather slowly or remained stationary until some movement caused them to disappear. Almost everywhere were multitudes of coarse or fine wrinkles. They were most marked about the knees, where they encircled the patellæ, partly arranged in semicircles, partly in elliptical forms. On standing the wrinkling became more striking. Similar but finer wrinkles occurred, as in my case, over the nates. While in a recumbent posture the color of the skin was brown from pigmentation, but on standing the veins rapidly filled and became distended, and the color of the surface became cyanotic. There was no perceptible difference in temperature from the normal. The hairs had mostly disappeared from the affected areas. That the sweat glands were involved was shown not only by the customary dryness of the affected skin, but by the fact that when pilocarpine was injected hypodermically in a sufficient dose to cause copious diaphoresis the atrophied portions of the skin remained as dry as before. Sensation was not impaired. A microscopic examination of an excised bit of the diseased integument showed that the adipose layer had entirely disappeared. There was a general atrophy of all the elements of the skin, the sweat glands were largely diminished, there was marked atrophy of the hair follicles, and the papillæ of the skin had disappeared. The connective tissue underneath the epithelial layer appeared swollen and infiltrated with cell nuclei. The nerves and blood-vessels were unchanged.

Idiopathic cutaneous atrophy of the extremities, such as in this case of Buchwald's, to which my case so closely corresponds, is undoubtedly rare. Several other cases, however, have been reported which are so nearly alike in their general characters as to warrant us in regarding Buchwald's atrophy as a peculiar variety of disease. Two cases that are apparently of this variety have been reported by Pospolow,* though in these the symmetry was less pronounced and the laxity of the skin (judging from a photograph that accompanies one of the cases)

* *Annales de Derm. et de la Syph.*, 1886, p. 505.

was somewhat greater than either in Buchwald's case or in my own. The photograph shows dependent folds of skin resembling dermatolysis. Also in Pospolow's cases the sweat glands were unaffected; but in the main the features corresponded to those of Buchwald's atrophy.

Touton * and Kristian Groen † have reported cases that are more typical. In Touton's case both upper and lower extremities were affected symmetrically. Groen's case, in the location and extent of the atrophy, corresponded almost precisely to my own case. The patient was a sailor, forty-seven years of age, who entered the hospital for atonic ulceration of the leg, and had apparently paid little attention to the atrophic condition, and of this no history was obtainable.

* *Deutsche med. Wochensch.*, 1886, p. 6.

† *Norsk Magazine.* Referred to in the *Lancet*, Nov. 28, 1891, p. 1238.